ORIGINAL ARTICLE

Deformity Correction, Surgical Stabilisation and Limb Length Equalisation in Patients with Fibrous Dysplasia: A 20-year Experience

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Abstract

Introduction: Fibrous dysplasia (FD) of bone can be present with pain, deformity and pathological fractures. Management is both medical and surgical. Little literature exists on the surgical management of both monostotic and polyostotic FD. We present our experience of limb reconstruction surgery in this pathological group of bone disease.

Materials and methods: A retrospective cohort of children who underwent limb reconstruction surgery at a single high-volume paediatric centre was identified from a prospective database. Case notes and radiographs were reviewed. Surgical techniques, outcomes and difficulties were explored.

Results: Twenty-one patients were identified aged between 7 and 13 at presentation to the limb reconstruction unit. Eleven were female, nine had McCune-Albright syndrome, seven had polyostotic FD and five had monostotic. Proximal femoral varus procurvatum deformity was the most common site requiring surgical intervention. The distal femur, tibia, humerus and forearm were also treated.

Methods include deformity correction with intramedullary fixation including endo-exo-endo techniques, elastic nailing, guided growth, circular fixator technique and fixator-assisted plating. Correction of deformity and leg length discrepancies was common.

The osteotomies went on to heal with no nonunions or delayed healing. We encountered secondary deformity at distal end of nails as the children grew as expected. These were managed with revision nailing techniques and in some cases external fixation. There was one implant failure, which did not require revision surgery.

Conclusion: The surgical management of pathological bone disease is challenging. Corrective osteotomies with intramedullary fixation can be very successful if appropriate limb reconstruction principles are adhered to. Deformity correction, guided growth and lengthening can all be successfully achieved in bone affected by FD. Polyostotic FD can be present with secondary deformities, and these can be difficult to manage.

Keywords: Fibrous dysplasia, Limb lengthening, McCune-Albright syndrome, Osteotomy.

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Introduction

Fibrous dysplasia (FD) is a rare, benign genetic condition of bone. Increased intracellular levels of cAMP in bone-forming cells result in immature connective tissue within the bone structure and poorly formed immature trabecular bone.[1,2] This presents the treating surgeon with a complex and challenging problem as the structural integrity of the skeleton is affected, leading to pain, pathological fractures and in some cases severe deformity.[3,4] FD can be present in numerous ways, with lesions in both upper and lower limb segments, spine and craniofacial area. FD presents as monostotic disease in approximately 80% of cases and with the remaining 20% of cases presenting as polyostotic disease;[4-6] aggressive polyostotic disease[7] is usually associated with McCune-Albright syndrome.[6,8] Management of FD requires a multidisciplinary approach, with involvement of endocrinologists, metabolic bone specialist and surgical specialties. Many patients with mild disease will not require surgical intervention.

Specific challenges are present when surgically treating a growing child who has FD, as whilst the child grows secondary deformities will likely present around any previous deformity corrections or fixations. Little literature currently exists to help guide management in skeletally immature patients, specifically with the use of modern implants such as rigid intramedullary nails and lengthening nails. A wide range of different surgical techniques is employed when addressing both upper and lower limb deformity corrections.

The aim of this study was to review the 20-year experience of the surgical management of FD in skeletally immature patients treated at a tertiary referral paediatric limb reconstruction unit within the United Kingdom (UK).

Materials and Methods

A retrospective analysis was performed to collect data from a prospective database on all consecutive patients with FD surgically

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treated at a tertiary referral paediatric hospital over a 20-year period. All case notes, operative notes and radiographic imaging were reviewed from time of presentation to latest follow-up or discharge from the limb reconstruction unit.

Data were collected to include patient demographics, specific diagnosis, limb segments affected, surgical techniques used, implants used, outcomes and complications.

We reviewed the presentation and management of secondary deformities that are specific to the growing child.

Results

Data were collected from 21 consecutive patients with FD requiring corrective surgery. Average age was 9 years (7–13) at the time of initial presentation to the limb reconstruction unit including 11 females and 10 males.

The majority, 16 patients, had polyostotic disease, with McCune-Albright syndrome being the most common diagnosis seen in 10 of these patients. Five patients treated had isolated monostotic disease.

Reasons for presentation to the limb reconstruction unit included pain, incidental findings on radiographs, deformity, pathological fractures or referrals from endocrine or metabolic bone services.

Five patients were tertiary referrals for the management of secondary deformities after initial fracture fixations or deformity corrections of the femora.

The lower limb segments required surgical intervention greater than the upper limb segments (Graph 1). However, surgery was performed either in isolation or combination on the proximal femur, distal femur, tibia, humerus and forearm. Proximal femoral deformity required the most reconstructive surgery, in 11 separate patients (Fig. 1).

Pathological Fracture

Pathological fractures were common, seen in both the femur and the tibia. Four patients presented following a low-energy trauma, falls from a standing height. A single patient presented following a high-energy trauma from a road traffic collision.

Different techniques were used in the fracture management, all required surgery. Intramedullary fixation was the preferred method of fixation (Figs 2A to C). Three patients with femoral fractures were treated with closed reduction, and intramedullary (IM) nailing, rigid locked paediatric nails were used when possible. One patient required elastic intramedullary nailing of their femoral fracture due to small size and low body weight.

One patient with a proximal femoral coxa vara deformity sustained a pathological fracture at the site of deformity; this was treated with acute corrective valgus osteotomy and IM nailing. This allowed fixation of the fracture and correction of the mechanical axis of the lower limb to allow the ideal mechanical environment for fracture healing.

One tibial fracture, sustained in a road traffic accident (RTA), was treated with external fixation, and fracture union was achieved with good alignment; however, the pathological nature of bone meant that a refracture occurred 6 months following frame removal. The subsequent fracture required IM nailing.

Deformity

Deformity in FD is common and was the main indication for surgery. Different techniques can be used to correct the mechanical axis including guided growth, epiphysiodesis and osteotomy with acute or gradual correction. A wide range of implants is available to achieve fixation following an osteotomy. It is the senior author's experience that IM fixation is the gold standard when managing children with FD; this allows for only acute corrections but protects and spans the length of the bone. The aim of deformity correction was to correct and maintain the mechanical axis of the lower limb throughout growth until skeletal maturity (Fig. 3).

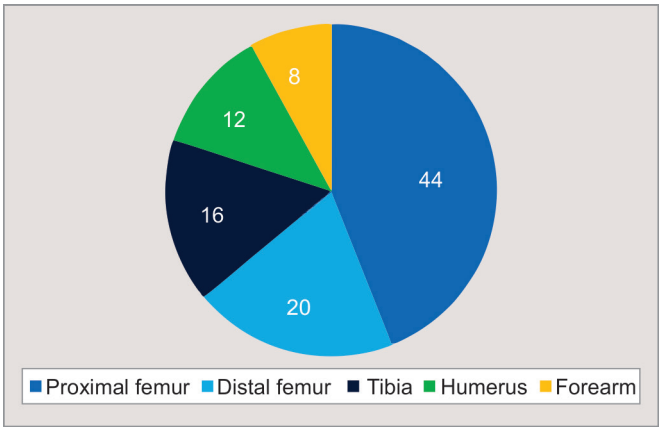
Lower Limb Deformity

Coronal plane alignment can be safely and effectively corrected by guided growth in patients with FD. Guided growth in the form of hemi-epiphysiodesis was used both in isolation and in combination with nailing techniques to correct the coronal alignment of the lower limb.

Proximal femoral coxa vara with or without concomitant procurvatum was the most common deformity. We performed 14 proximal femoral corrections.

Techniques used included:

- Valgus realignment osteotomy with paediatric locking IM nails,
- Valgus osteotomy with growing rods and tension band construct,



Graph 1: Illustration of the different limb segments affected which required reconstructive surgery. The proximal femur was the most commonly affected limb segment

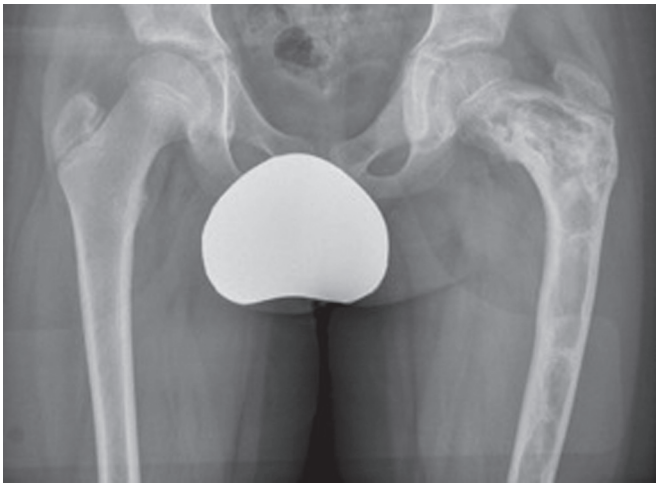
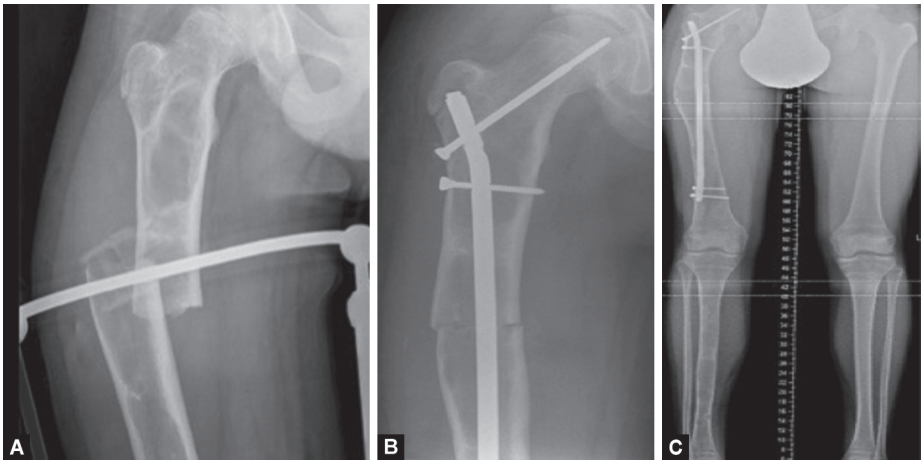


Fig. 1: Radiograph illustrating left-sided coxa vara proximal femoral deformity



Figs 2A to C: Radiographs illustrating a pathological fracture of the proximal femur. Treated with closed reduction and locked rigid lateral entry paediatric IM nailing

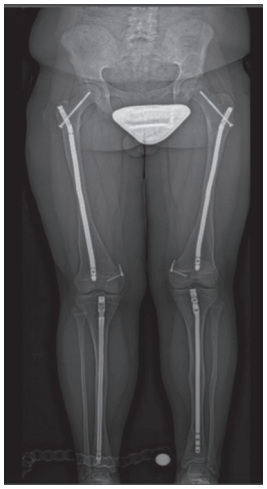
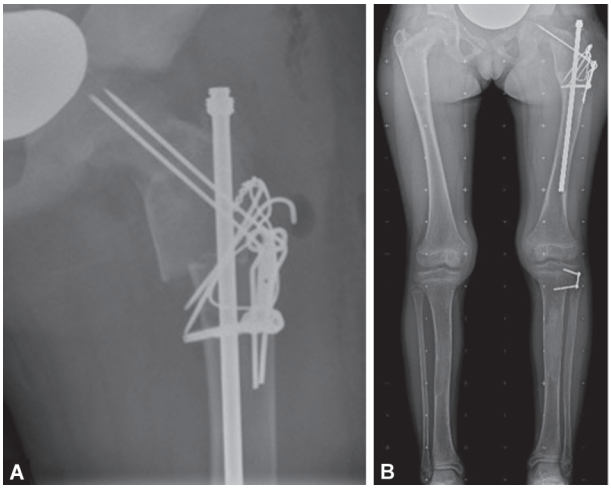


Fig. 3: Mechanical axis radiograph illustrating four-segment deformity correction with IM fixation and guided growth to achieve the desired alignment at skeletal maturity



Figs 4A and B: Radiographs illustrating a tension band wire technique utilised in a complex deformity correction of the proximal femur. When the femoral neck is not accessible with a cephalomedullary screw, tension band wire constructs can be used instead.

- Osteotomy with lengthening nails to correct leg length discrepancy (LLD),
- End-exo-endo nailing technique (Figs. 4A and B).

Two cases of distal femoral deformities were treated with osteotomy and retrograde rigid nailing.

Upper Limb Deformity

Upper limb deformity less frequently required surgical correction. We treated five humeri and three forearms for deformity correction. Similar principles were utilised when managing upper limb deformities, osteotomies and IM nailing to correct alignment.

Techniques used included:

- Osteotomy and antegrade nailing for humeri (Figs. 5A and B),
- Osteotomy and retrograde nailing for humeri,
- Multiple osteotomies and flexible nailing for the forearm (Fig. 6).

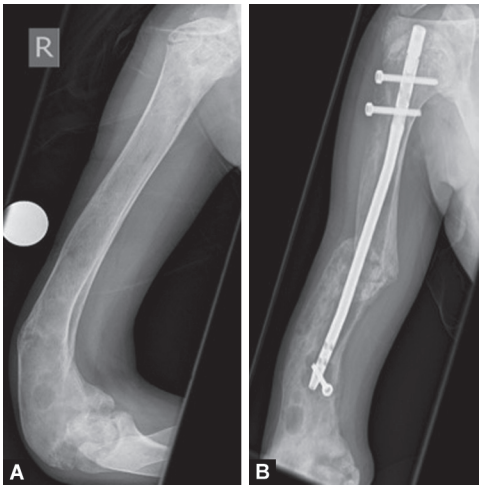
Secondary Deformity

Secondary deformity is common when managing FD in skeletally immature patients. It is seen following both fracture fixation and deformity correction surgery. It is recommended that patients are followed up until skeletal maturity and assessed both clinically and radiologically with mechanical axis radiographs. We observed that 13/16 (81%) of patients with polyostotic disease and 3/5 (60%) patients with monostotic disease required revision surgery for secondary deformities.

Patients who were treated at a young age with flexible nails were revised to modern paediatric locking nails when able during revision surgery.

Techniques used included:

- Revision nailing with corrective osteotomy and
- Guided growth (hemi-epiphysiodesis).



Figs 5A and B: Deformity correction of the humerus for impending pathological fracture. Both antegrade and retrograde nailing can be performed

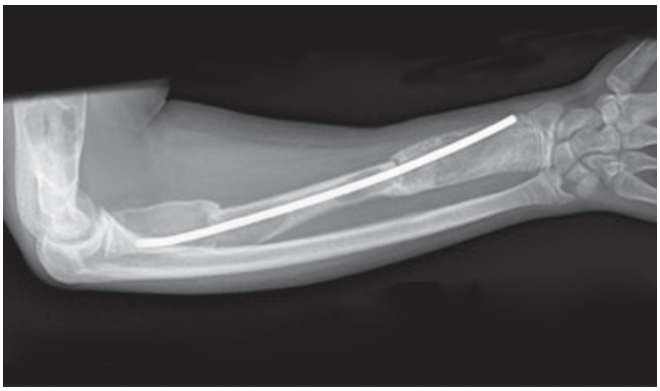


Fig. 6: Radiograph illustration of a double-level osteotomy required in the radius with IM flexible nail fixation

Complications

Although FD is a challenging condition to manage surgically, complications were generally low. Secondary deformity was common; however, this was expected with growth.

There was a single implant failure, in which there was a fracture of the proximal femoral nail that required revision surgery to a larger-diameter nail.

We did not observe any deep infections or nonunions in either the fracture or deformity correction group.

Leg length discrepancy (LLD) was common and reported in nine patients. This was often well tolerated and treated only with orthotics for a shoe raise. Three patients had significant LLD and warranted surgical correction. One patient underwent surgical growth arrest with a drill epiphysiodesis, and two patients required limb lengthening.

Symmetrical leg lengths were achieved by controlled lengthening of the femur. A magnetic lengthening nail, PRECICE 2 (P2 nail system), was used in both antegrade and retrograde fashion. We did not observe any complications with bone regeneration and FD demonstrated not to be a contraindication to lengthening.

Discussion

Fibrous dysplasia of bone is a challenging condition to surgically manage, particularly polyostotic disease in a growing child. There is limited literature to help guide decision-making, likely due to the low number of cases encountered. In this retrospective cohort, we have been able to highlight different techniques that can be employed to manage pathological fracture, deformity, secondary deformity and limb length discrepancies in this patient cohort. The experience of Stanton et al. in the management of FD is similar to ours; intramedullary fixation was preferred over plate and screws. They also encountered severe deformities and secondary deformities in the growing child, many of which were managed in a similar fashion.[9]

Previous studies have discussed the use of bone grafting for ablation of FD; this was first described in 1987 by Enneking and Gearen.[10] It has been our experience that bone grafting for ablation is not required in FD.[11] We also do not advocate excision or curettage of the involved segment. We did not experience any nonunions in either the fracture or the deformity correction group, even when the osteotomy was performed at this site of the dysplastic bone.

Proximal femoral deformities are the most common deformity encounters and may present a significant surgical challenge.[12-14] Coxa vara deformity predominates and the ideal method of managing this deformity is with a valgus osteotomy and IM nail to correct the mechanical axis. Some deformities are so severe that standard devices do not fit the proximal femoral anatomy. Different techniques can be used in this scenario. Plate and screw fixation has previously postulated to be the treatment of choice due to the challenges involved in passing an IM device.[15] We believe that IM fixation is advantageous as it protects the length of the bone and provides the optimal mechanical environment for healing. If the femoral neck is not accessible, tension band constructs can be made to enhance the fixation. If the proximal femur does not allow for the passage of a rigid IM nail, endo-exo-endo nailing techniques can be used.

Revision surgery is common in the growing child seen in 81% of patients treated with polyostotic disease, which is inevitable and is not considered as a complication of surgery. The gold standard for the treatment of secondary deformity is revision nailing with an osteotomy at the site of the new deformity.

Limb length discrepancy is common. This most commonly can be managed with orthotics. However, if surgery is warranted, growth can be stopped surgically by drill epiphysiodesis of the longer limb or bone can be lengthened with modern intramedullary lengthening nails. We did not experience any problems with bone regeneration or complications with lengthening in children with FD.

A limitation of this study is that some of the surgical techniques presented do not have a robust evidence base when used in the treatment of FD; this is due to the paucity of evidence currently available for surgeons who manage patients with FD. We hope this study provides surgeons with suitable evidence that osteotomy, IM fixation, guided growth and limb lengthening can be safely and effectively utilised to correct deformity and limb length inequalities in patients with FD.

Conclusion

The surgical management of FD in the growing child consists of the analysis of deformities, close monitoring, surgical interventions

and long-term follow-up. Indications for surgery are pathological fractures and severe deformities, most commonly of the proximal femur. Intramedullary fixation of fractures is preferred whenever possible. When performing deformity corrections, the surgeon must be familiar with many different implants and techniques to correct the mechanical axis. Limb length equalisation can successfully be achieved in patients with FD.

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